- 1 A substantially pure mannin-binding lectin 2 associated serine protease-2 (MASP-2) polypeptide.
- 2. The polypeptide of claim 1, said polypeptide being capable of associating with mannan-binding lectin (MBL).
- 3. The polypeptide of claim 1, said polypeptide being conjugated to a label or toxin.

4. A polypeptide containing the sequence identified as SEQ ID NO. 1.

- 5. A polypeptide according to claim 4 with a molecular mass of 20K.
- 6. A polypeptide with a molecular mass of 52K and containing the sequence identified as SEQ ID NO 1.
- 7. The polypeptide of claim 1, said polypeptide having serine protease activity.
- 8. A polypeptide of claim 1, said polypeptide being capable of MASP-2 activity in an *in vitro* assay for MBLectin complement pathway function.
- 9. A polypeptide according to claim 1, said polypeptide being capable of competitively inhibiting MASP-2 serine protease activity.

10. A polypeptide according to claim 1 comprising a 2 fragment of the polypeptide of SEQ ID NO:2, said polypeptide 3 being a competitive inhibitor of complexing of MBL/MASP-2.

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- 11. A polypeptide according to claim 5 or claim 6, said polypeptide having the amino acid sequence of SEQ ID NO:2.
- 1 12. A compound capable of competitively inhibiting/
 2 serine protease activity of MASP-2 or a fragment thereof.
 - 13. An isolated nucleic acid molecule of claim the molecule comprising a nucleotide sequence encoding a polypeptide having sequence that is at least 85% identical to the sequence of SEQ ID NO:1 or 2.
- 1 14. An isolated nucleic acid sequence encoding a 2 mannan-binding lectin associated serine protease-2 (MASP-2) 3 polypeptide according to claim 1.
- 1 15. A nucleic acid vector comprising the nucleic 2 acid molecule of claim 14.
- 1 16. The nucleic acid vector of claim 15 wherein 2 said vector is an expression vector.
- 1 The vector of claim 16, further comprising a regulatory element.
- 1 18. An antibody produced by administering an MASP-2 polypeptide according to claim 1 to an antibody producing 3 animal.

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1	19. An antibody that selectively binds to MASE-2.
1	20. The antibody of claim 18 or claim 19, wherein said antibody is a monoclomal antibody.
1 2	21. The antibody of claim 18 or 19, said antibody being coupled to a compound comprising a detectable marker.
5 (1) 1 '	22. A pharmaceutical composition comprising the polypeptide of claim 1 or the antibody of claims 18 or 19.
1	23. A method for detecting mannin-binding lectin
2	associated serine protease-2 (MASP-2), said method
3	comprising:
4	(a) obtaining a biological sample;
5	(b) contacting said biological sample with a MASP-2
6	polypeptide specific binding partner that specifically binds
7	MASP-2; and
8	(c) detecting said complexes, if any, as an
9	indication of the presence of magnin-binding lectin
10	associated serine protease-2 in said sample.
1	24. A method according to claim 23, in which the
2	specific binding partner is an antibody.
1	25. A method for detecting MASP-2, said method
2	comprising an assay for MASP-2 complement fixing activity.
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1	26. The methods of claims 23 or 24 for quantitative

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assay of MASP-2 or MASP-2 activity in biological samples.

- 27. A method for detecting MASP-2 nucleic acid expression, comprising detecting RNA having a sequence encoding a MASP-2 polypeptide by mixing the sample with a nucleic acid probe that specifically hybridizes under stringent conditions to the nucleic acid of claim 13 or 14.
- 28. A method for treating patients deficient in MASP-2 by administering to the patient the peptide of claim 1.
- 29. A method for treating patients deficient in MASP-2 by administering to the patient nucleic acid according to claim 13 or 14.
- 30. A method for inhibiting the activity of MASP-2 by administering to the subject a compound that inhibits expression or activity of MASP 2.
- 1 31. The method of claim 27 in which the compound is 2 a MASP-2 anti-sense nucleic acid sequence.
- 32. The method of claim 30 comprising administering a compound that inhibits complexing of MBL and MASP-2.
- 33. An assay for polymorphisms in the nucleic acid sequence encoding MASP-2.

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- and a method of detecting the presence of MASP-2 encoding nucleic acid in a sample, comprising mixing the sample with at least one nucleic acid probe capable of forming a complex with MASP-2-encoding nucleic acid under stringent conditions, and determining whether the probe is bound to sample nucleic acid.
- 35. A nucleic acid probe capable of forming a complex with MASP-2-encoding nucleic acid under stringent conditions.
 - 36. An assay for polymorphisms in the polypeptide sequence comprising MASP-2 or its precursor.
 - 37. A method for diagnosing a disorder associated with aberrant expression of MASP 2, comprising obtaining a biological sample from a patient and measuring MASP-2 expression in said biological sample, wherein increased or decreased MASP-2 expression in said biological sample compared to a control indicates that said patient suffers from a disorder associated with aberrant expression of MASP-2.
 - 38. A method for diagnosing a disorder associated with aberrant activity of MASP-2, comprising obtaining a biological sample from a patient and measuring MASP-2 activity in said biological sample, wherein increased or decreased MASP-2 activity in said biological sample compared to a control indicates that said patient suffers from a disorder associated with aberrant activity of MASP-2.

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:1 39. A method of assaying for activity MBL-complexed MASP, the method comprising 2 providing a sample to be assayed and substantially 3 reducing any artifact resulting from activation of the 4 classical complement fixing bathway by conducting the assay 5 in the presence of an ionic strength high enough to 6 effectively reduce activation of the classical complement 7 fixing pathway but not so high as to substantially interfere 8 with activity of MBL-complexed MASP.

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